

Occipital Cephalocele With Brain Stem Herniation

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Citation

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Abstract

We report an interesting and unique case of occipital cephalocele with herniation of the brain stem and cerebellar peduncles. Interestingly the patient had no neurological deficits.

INTRODUCTION

Occipital cephaloceles refer to a defect in the skull and dura with extracranial herniation of intracranial structures (^{1, 2}) and are rare congenital malformations. They can occur in isolation or with various syndromes. We describe heretofore undescribed imaging features, in a neurologically asymptomatic child.

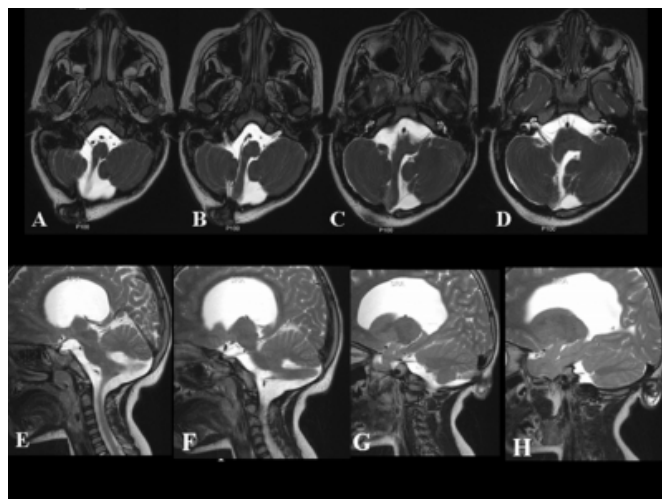
CASE REPORT

A 5-year-old male child, born 6 weeks premature, of a non-consanguineous parentage, presented with a swelling over the right occipital region since birth. There was no history of perinatal head trauma. The swelling was soft to firm in consistency, non pulsatile and had no impulse on crying. There was no discharge or leak from the swelling. There was no increase in head circumference and the lesion remained static in size over these years. Developmental milestones were normal.

Magnetic resonance imaging (MRI) was done on a 1.5 T scanner, and images obtained in various orthogonal planes with T1 and T2 weighting. In addition, high-resolution thin T2 weighted axial images at the level of the occipital swelling were also obtained. The MRI revealed kinking of the brainstem at the pontomedullary junction with a band of tissue extending posteriorly across the posterior fossa to end in the cephalocele sac. This band was a continuation of the right half of the lower part of the superior cerebellar peduncle, the entire middle and inferior cerebellar peduncle and part of the superior medulla. CSF extended around the band upto the inner occipital margin {Figure 1(A-H)}.

Figure 1

Figure 1(A-H): Axial T2 weighted FSE images (A-D) of the posterior fossa reveal the abnormal band of tissue surrounded by CSF extending from the brainstem to the occipital cephalocele. Note the stretching of right VII- VIII nerve complex (D) and the right posterior inferior cerebellar artery (B).



Sagittal T2 weighted FSE images (E-H) shows kinking of the brainstem and the abnormal band arising from the brainstem opposite to the kink. The cephalocele is located in the lower occipital region.

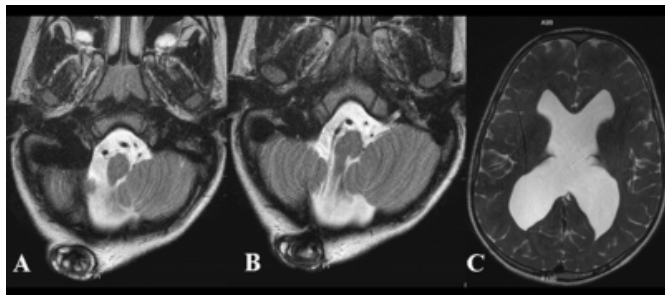
The band continued posteriorly into the cephalocele sac, which contained T2 mixed hyperintense tissue – likely dysplastic

{Figure 2 (A, B)}. The right cerebellar hemisphere was hypoplastic. The right VII- VIII nerve complex was stretched, as was the right posterior inferior cerebellar artery. The vermis was absent and there was abnormal communication between the fourth ventricle and cisterna

magna. The supra tentorial compartment revealed a thin corpus callosum with dilated lateral ventricles and absent septum pellucidum {(figure 2C)}.

Figure 2

Figure 2 (A-B): Thin axial T2 weighted FSE images shows mixed intensity tissue within the cephalocele (C) The supratentorial axial T2 weighted FSE image shows the dilatation of lateral ventricles and absence of septum pellucidum .



DISCUSSION

Cephalocele refers to a defect in the skull and dura with extracranial herniation of intracranial structures.

Cephaloceles occur approximately in 1- 3 in 10,000 live births. Osborn states the incidence of occipital cephaloceles to be 70 – 90% of all cephaloceles. (3). The development of a cephalocele has been attributed to various mechanisms by different authors .Van Allen attributes development of cephaloceles to failure of primary neurulation (4). However Gluckman et al state that cephaloceles are a result of a post neurulation event in which the brain tissue herniates through a defect in the mesenchyme that is to become the cranium and dura (5).

Antenatal diagnosis is possible with ultrasound .CT demonstrates the osseous defect. However; MR plays a definitive role in demonstrating the contents of the sac, while MR venogram demonstrates the associated venous sinus abnormalities.

Based on its contents, cephaloceles are classified into four types, meningoencephaloceles, meningocele, atretic cephaloceles & glioceles. Based on the bone defects through which they course, they are classified as occipitocervical, occipital, parietal, frontal, temporal, frontoethmoidal, sphenomaxillary, sphenoorbital, nasopharyngeal and lateral. (6).

Occipital cephaloceles originate between the foramen magnum and the lambda (7). They contain dysplastic and gliotic brain tissue within. It may be seen in isolation or as a

part of a syndrome. Associated anomalies include absent corpus callosum, orofacial clefting, craniostenosis, Dandy-Walker defect, Arnold-Chiari defect, ectrodactyly, hemifacial microsomia, hypothalamic-pituitary dysfunction, Klippel-Feil anomaly, iniencephaly, and myelomeningocele (8).

Our patient presents unique features as regards the clinical manifestations and the imaging findings. The imaging features of herniation of the cerebellar peduncles through a high occipital osseous defect into the encephalocele sac are to the best of our knowledge not described in the literature as yet. The interesting clinical manifestation was that in spite of significant imaging findings, the patient had no neurological deficits.

To conclude, unusual imaging findings as seen in this asymptomatic child highlight the need for doing appropriate imaging techniques for diagnosis and further management in children with cephaloceles. .

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